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Diagnostic dilemma of a complex tumor-hemangiopericytoma/ solitary fibrous tumour or nodular hidradenoma of thigh - A unique case report

Pooja Jha^{1*}, Samarth Shukla², Sunita Vagha³, Ravindra P Kadu², Sourya Acharya⁴

ABSTRACT

Hemangiopericytoma is described as a rare variety of tumors originating from the unrestricted or uncontrolled proliferation of pericytes. It is believed that these rare tumors arise from the vascular cells called Zimmerman pericytes which are present in the entire spiral body involving the capillaries and post-capillary venules. These painless, slowly growing tumors are liable to develop in the subcutaneous tissue and skeletal muscle. Hemangiopericytomas more commonly affect the lower extremities and may also affect the head and neck region. Clinically, it can affect any age but the increased incidence has been noted in adults in the fifth decade of life without any gender predilection. We report a case of hemangiopericytoma of the left lower extremity i.e., in the thigh in a 35-year-old male patient who came to our hospital at J.N.M.C., Sawangi (M), Wardha with the chief complaints of a painless mass in the left thigh which clinically looked like a sebaceous cyst. Thereafter, tumor excision was performed and the sample was sent for HPE. The histopathological examination of the tumor tissue showed features that were highly suggestive of hemangiopericytoma. However, interestingly the tumor was diagnosed as Nodular Hidradenoma, a benign adnexal tumor on immunohistochemistry. Nodular hidradenoma is a rare non-malignant tumor of adnexa that originates from the distal excretory duct of eccrine or apocrine sweat glands, which is rare, considering all the adnexal tumors.

Keywords: Hemangiopericytoma, pericytes, benign mesenchymal tumors, solitary fibrous tumors, nodular hidradenoma

1. BACKGROUND

Hemangiopericytoma was first described by Stout and Murray in 1942 as rare neoplasms arising from pericytes surrounding blood vessels. 691 cases were of

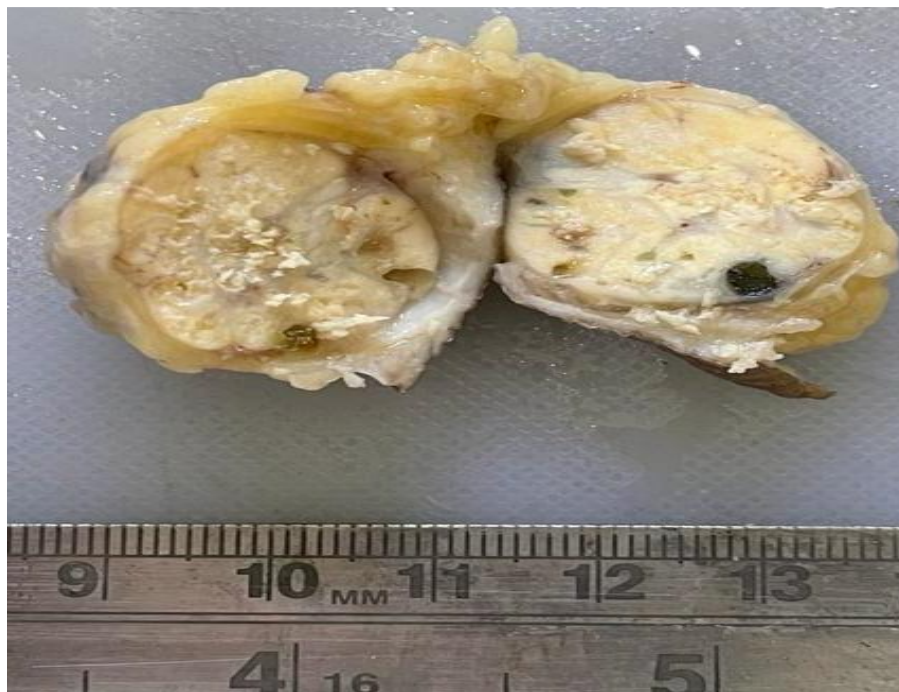
vascular tumors, and only nine of them were hemangiopericytomas (Stout et al., 1942; Tauro et al., 2013). However, the tumor received widespread recognition after 1949 when Stout reported 25 additional cases. These solitary fibrous tumors are benign mesenchymal tumors that account for about upto 5 % of all soft tissue sarcomas and rarely around 1% of all vascular tumors (Tauro et al., 2012). The underlying etiology or mechanisms are still not clear; also the diagnosis and management guidelines for hemangiopericytoma are not well established yet.

Amongst all the benign adnexal tumors i.e., syringoma, poroma, cylindroma, spiradenoma hidradenoma papilliferum, hidradenoma etc., we will discuss about the features of one of the rare adnexal tumors i.e., nodular hidradenoma. It was previously assumed that it arises from the eccrine glands but now it is suggested that this tumor most often originates from the apocrine glands (Benkirane et al., 2020). The term nodular hidradenoma or eccrine acrospiroma was coined by Johnson & Helwig (Shruthi et al., 2021). The first case of nodular hidradenoma was described in the literature by Delacretaz and Leresche (Shruthi et al., 2021). These are sweat gland neoplasms and are extremely unique. They are benign and can be of either eccrine or apocrine origin, the latter being more common.

2. CASE HISTORY

A 35-year-old male patient first presented to the surgery outpatient department of our hospital J.N.M.C., Sawangi (M), Wardha with chief complaints of pain and swelling in the left thigh for about 8-10 months. The swelling was gradationally growing in size and was painless. On clinical examination, the mass was about 6 x 5 x 2.5 cm and was firm and non-tender, overlying skin was normal, was not mobile, and not fixed to the overlying structures. There was no local rise in temperature. Family history and personal history were non-contributory. No abnormalities were detected on general and systemic examinations of the patient.

The plain pelvic radiographs with the left femoral bone showed a soft tissue lesion over the upper third region of the left thigh with osteolytic damage of the left femur in the proximal one third area. The subject also gave a history of a fall sustaining an injury to the left hip 3 years back. MRI was suggestive of a single well-defined heterogeneous intensity mass in the anterolateral aspect of the left thigh with some calcification areas suspicious of hemangiopericytoma. Tumor was excised and the specimen was sent & examined in the Department of Pathology. Grossly, the tumor was a solitary mass at the anterolateral aspect of the left thigh. It was fibrous and was firm to cut and was well-circumscribed and encapsulated. It had been excised along with the surrounding skin and fat. The external surface was nodular and smooth. On the cut section, whitish, greyish solid cystic areas and a few hemorrhagic areas were also noted (Figure 1 & 2).



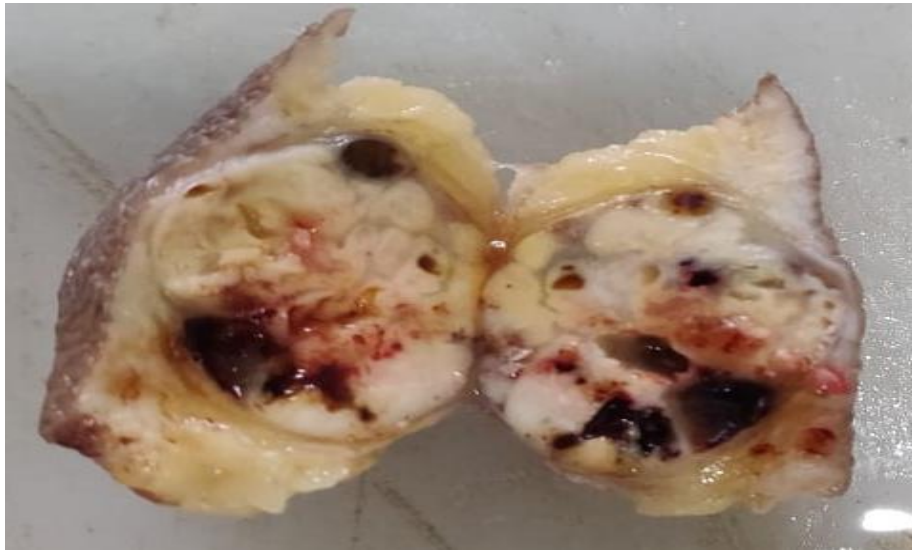


Figure 1 & 2 Grossly, the tumor was a solitary mass at the anterolateral aspect of the left thigh. It was fibrous and was firm to cut and was well-circumscribed and encapsulated. It had been excised along with the surrounding skin and fat. The external surface was nodular and smooth. On the cut section, whitish, greyish solid cystic areas and a few hemorrhagic areas were also noted.

Microscopically, the histopathological features suggestive of hemangiopericytoma were the tumor cells that appeared as ovoid to fusiform spindle cells with smothered borders arranged in a few short, ill-defined fascicles (Figure 3). Few dilated branched, hyalinized staghorn-like vasculature were seen. Fibroblast-like cells within a collagenous stroma, necrosis, and atypia were not present (Figure 4). The histopathological features suggestive of nodular hidradenoma noticed were at places, and epithelial cells in solid components showed vague nodular arrangement. The solid component was observed to have two cell types: polyhedral cells with basophilic cytoplasm and glycogen containing clear cells with an eccentric round nucleus. Foci of squamous differentiation were seen. Mitosis was sparse (Figure 5).

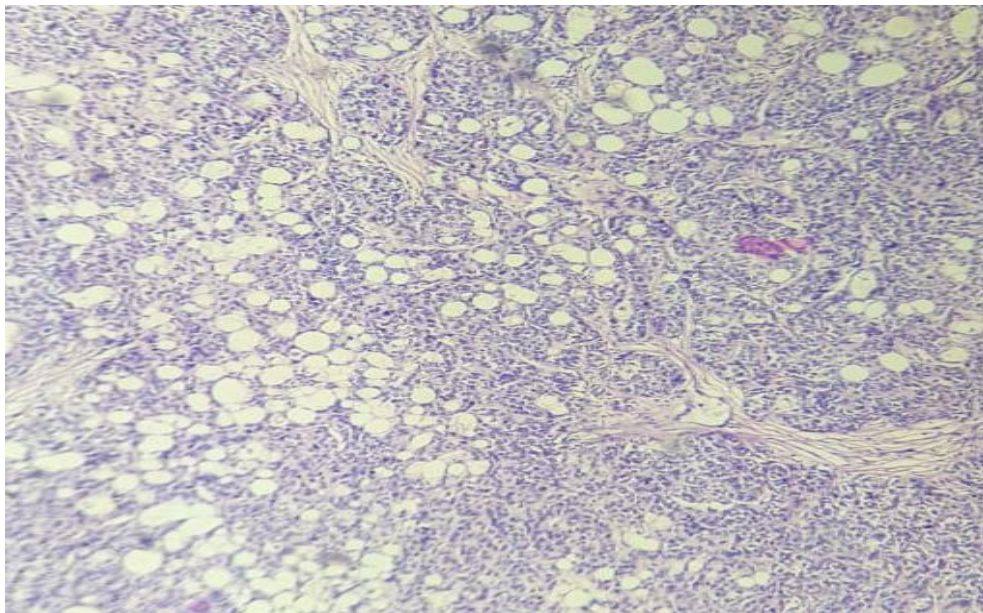


Figure 3 Microscopic H and E staining 10 x magnification

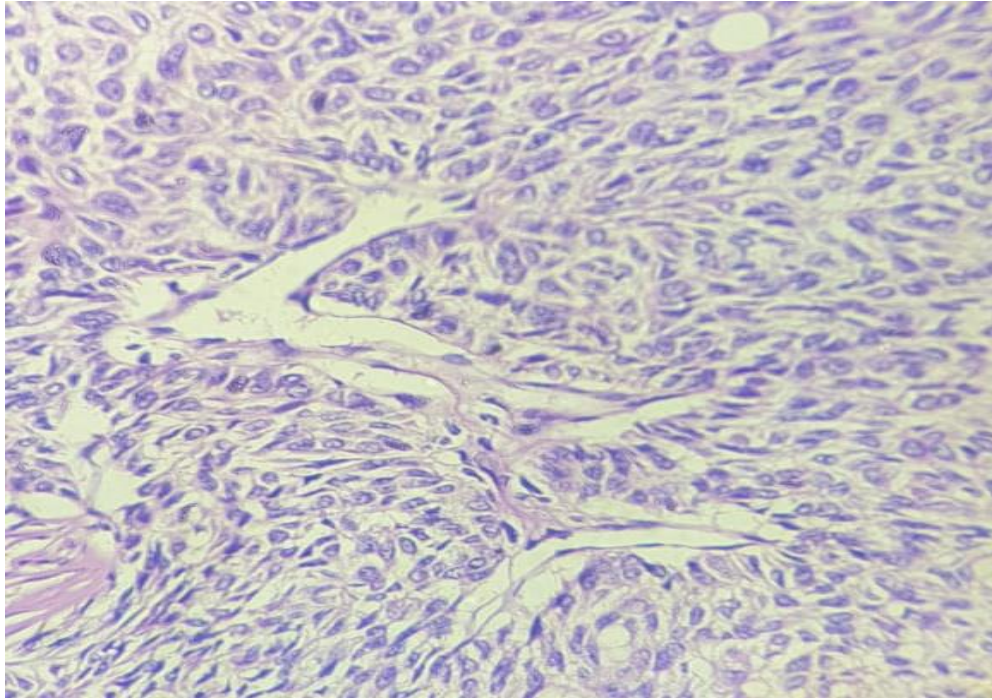


Figure 4 Microscopic H and E staining 40 x magnification

The tumor more likely appeared to be hemangiopericytoma on microscopy as many cells appeared to be ovoid to spindle cells with few dilated branched hyalinized staghorn like vasculature but the polyhedral cells with basophilic cytoplasm and glycogen containing clear cells with an eccentric round nucleus created the plight under the lens. The microscopic dilemma invited the need of performing immunohistochemistry for this patient. On IHC, tumor cells showed diffuse immunoreactivity for AE1/AE3, moderate diffuse immunoreactivity for CK5/6, and focal immunoreactivity for SMA. Tumor cells were immune-negative for CD 34, CD 31, MDM 2, Beta Catenin, S-100, and Desmin. MIB-1 labelling index was approximately 14%. This immune profile and histomorphology were suggestive of Nodular Hidradenoma (Figure 6 & 7).

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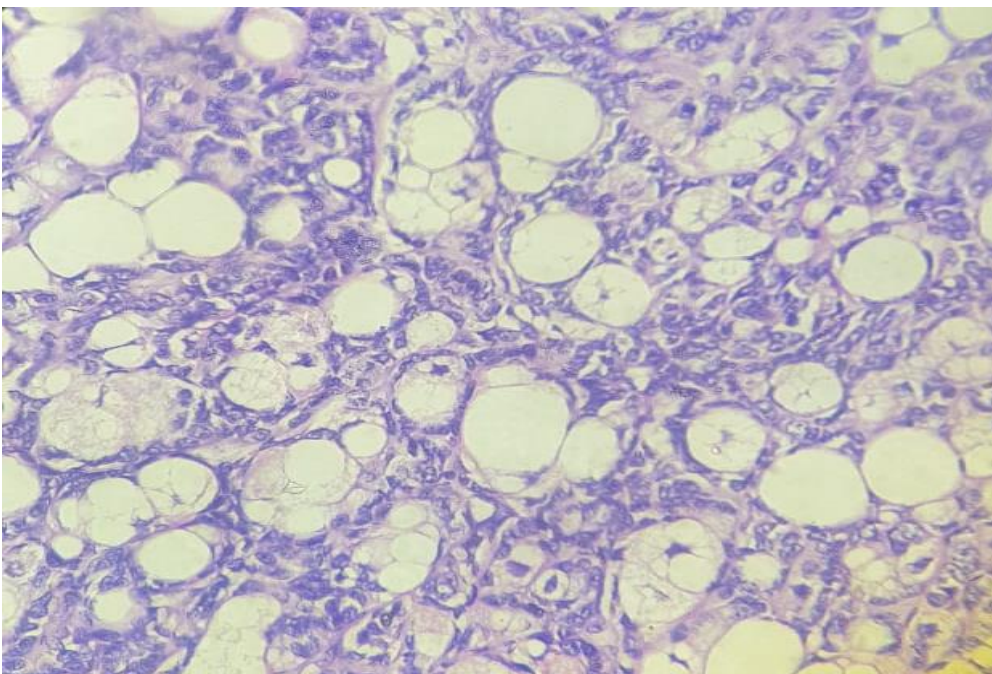


Figure 5 Microscopic H and E staining 40 x magnification

Histopathology showed well-defined aggregates of cells in the dermis comprising clear cells and polygonal with abundant glycogen, whorls of squamoid cells with eosinophilic cytoplasm and mucinous cells. Nodules interlaced with dermal ducts showing two layers of cells and glandular areas and reported as Nodular Hidradenoma.

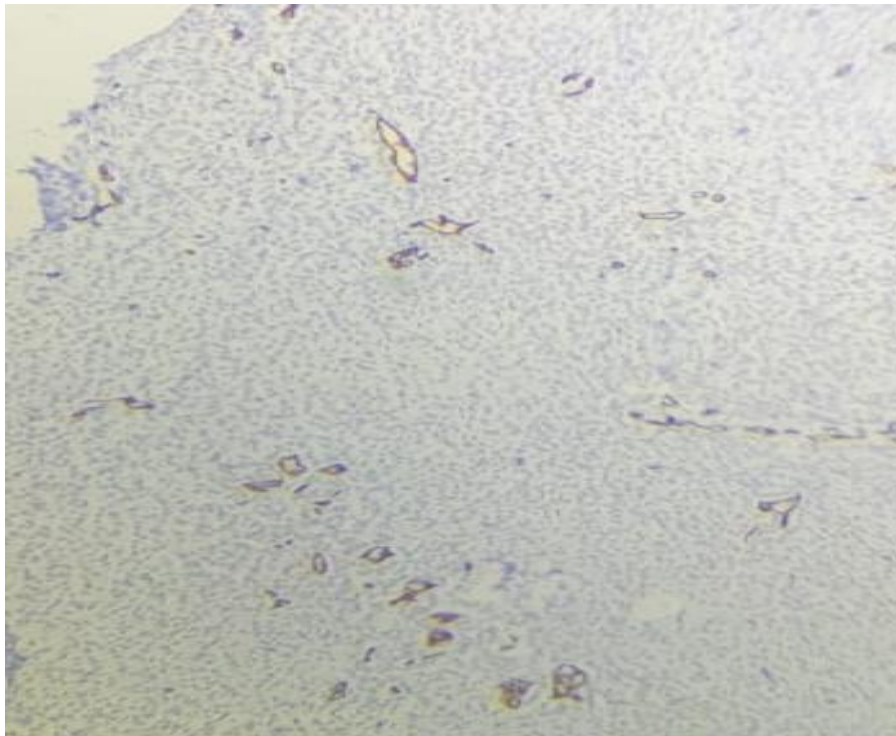


Figure 6 IHC staining 10 x magnification

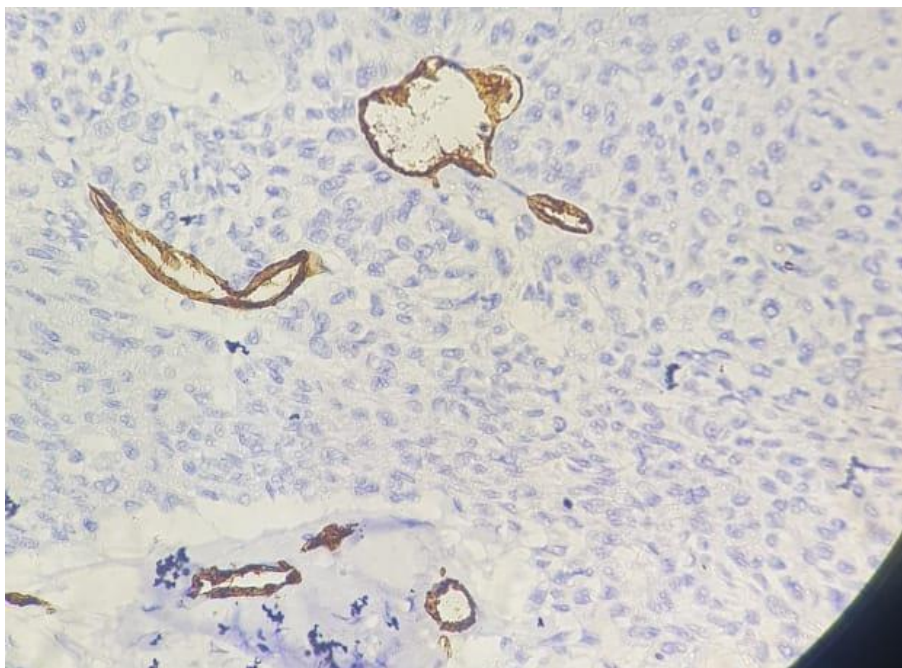


Figure 7 IHC staining 40 x magnification

On IHC, Tumor cells showed diffuse immunoreactivity for AE1/AE3, moderate diffuse immunoreactivity for CK5/6, and focal immunoreactivity for SMA suggestive of nodular hidradenoma.

3. DISCUSSION

In 1942, hemangiopericytoma was first stated by Stout and Murray, is a relatively rare mesenchymal tumor observed most commonly during the fifth decade of adult life and is uncommon in children. It arises from the uninhibited propagation of pericytes with the potential for recurrence and metastasis. It can occur at any anatomical site but mainly involve the extremities, abdominal cavity, pelvis, retroperitoneum, head & neck, and trunk. It has been noticed that some patients remain asymptomatic until the disease progresses while the majority of patients present with pain and mass-related symptoms, which include a local rise in skin temperature, retention of urine, and constipation. Also, vascular disease-related symptoms may be seen in a few patients.

The etiology for hemangiopericytoma/SFT is still not clear and was initially considered to be separate entities, but the pathophysiology underlying the formation of both the tumors i.e., the identification of NAB2-STAT6 fusion and its nuclear relocation as a definitive molecular alteration has led to consider them as a single disease entity (Fritchie et al., 2019). Varied paraneoplastic syndromes were observed with hemangiopericytoma, including hypoglycemia, and hypophosphataemic osteomalacia (Mane et al., 2015; Atalabi et al., 2010; Karcioğlu et al., 1997; Grant et al., 1982; Taylor et al., 1988), and is a highly vascular tumor, clinically significant arteriovenous shunting can be seen occasionally. Hemangiopericytoma/SFT, especially when occurs in the thigh, must be distinguished from the more common sarcomas occur at the same site. As mentioned earlier, the tumor showed microscopic features of both SFT/Hemangiopericytoma and Nodular Hidradenoma, to rule out the confusion and to get a confirmatory diagnosis, immunohistochemistry was performed which is considered to have excellent sensitivity and specificity for histopathological diagnosis. The tumor showed features of both SFT/Hemangiopericytoma and Nodular Hidradenoma, on IHC the results were in the favour of Nodular Hidradenoma.

Nodular hidradenoma is a very infrequent benign adnexal tumor of the distant part of the sweat glands found usually in adult females in the fifth decennary of life. Unlike in our study, the patient is a 35-year-old male. It was previously assumed that it arises from the eccrine glands but now it is suggested that this tumor most often originates from the apocrine glands. These are solitary lesions mostly expected to be present on the scalp, face, or anterior trunk but here in our study, the tumor is located on the anterior aspect of the left thigh which is a rare case scenario. Sweat gland neoplasms are extremely unusual, allocated as benign and malignant. The benign tumors have been further sub-divided as nodular, apocrine, solid-cystic, and clear cells based on their histopathological pattern. The cancerous form is classified into two groups. The first group comprises malignant tumors that closely mimic their benign counterparts while the second group of tumors does not have a benign counterpart.

Clinically the tumor usually presents as an asymptomatic, solitary nodule, whose size varies from 0.5 to 6 cm. occasionally discoloration and surface erosions or ulceration may be observed. It is a slow-growing tumor and rapid growth may be due to trauma, hemorrhage, or a malignant change. Histopathology shows the solid areas which contain two types of cell population: polyhedral cells with basophilic cytoplasm and pale or clear cells carrying glycogen with clear cytoplasm and a round nucleus seen in our case. The precise frequentness of Nodular hidradenoma and its transformation peril into malignant tumors is unspecified. Malignant hidradenocarcinoma has rarely been reported. A sweat gland carcinoma often imitates the benign hidradenoma and customarily does not have any specific nuclear changes, hence biopsy is considered to ensure the tumor is non-malignant or malignant.

The current treatment followed for both hemangiopericytoma/SFT and Nodular Hidradenoma is wide surgical abscission with at least 2 cm of clear margins for both primary disease and recurrences. In Espat's report, patients who underwent tumor resection comprehensively showed a survival rate of 100% at about five years (Espat et al., 2002). Some authors have mentioned the postoperative complications related to surgical excision however, preoperative afferent ligation of vessels or vascular embolization might help to lessen the peril of operative hemorrhage. Both chemotherapy and radiotherapy seem effectual and are endorsed in all patients with insufficient resection and /or large, locally penetrating tumors. In our case, the patient was managed with complete tumor abscission, the tissue was sent for histopathological examination.

Final diagnosis

Even though the tumor seemed to have histopathological features similar to hemangiopericytoma/SFT and Nodular Hidradenoma, the diagnosis was confirmed as Nodular Hidradenoma on immunohistochemistry.

4. CONCLUSION

This case report showcases a very interesting scenario. Nodular hidradenoma is a very infrequent benign adnexal tumor of the distant part of the sweat glands found usually in adult females in the fifth decennary of life whereas, in our case the patient is a 35-year-old male. These are solitary lesions are mostly expected on the scalp, face, or anterior trunk but here in our study, the tumor is

located on the anterior aspect of the left thigh which is a rare presentation. Grossly and microscopically the tumor showed the histopathological features of both hemangiopericytoma/SFT and nodular hidradenoma. This microscopic confusion insisted the need for immunohistochemistry. The final diagnosis of the tumor was proven on Immunohistochemistry as nodular hidradenoma.

Although recent advances have helped and improved the diagnostic accuracy of SFT/HPC, the association of the NAB2-STAT6 fusion status with phenotype and prognosis of the disease remains unclear. Even the prognostic factors have not been established yet. This case report helps us to understand the current knowledge on the clinical, histological, and molecular characteristics of SFT/HPC. We also understood that in the case of nodular hidradenoma, clinical qualm is necessary and excisional biopsy could be the elected initial investigation and treatment of solitary nodular hidradenoma. Discrete adnexal tumors may have identical clinical features hence histopathological examination of the tumor is mandatory to diagnose whether it is non-malignant or malignant with immunohistochemistry being the final diagnostic tool.

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Authors' contribution

Dr. Samarth Shukla, Dr. Sunita Vagha: conceptualization and design; Dr. Samarth Shukla, Dr. Pooja Jha: Investigations, observation and analysis; Dr. Samarth Shukla, Dr. Sourya Acharya, Dr. Pooja Jha: draft preparation; Dr. Samarth Shukla, Dr. Pooja Jha, Dr. Ravindra P. Kadu: review and editing; Dr. Samarth Shukla: supervision. All the authors have read and agreed to the final manuscript.

Informed consent

Written & oral informed consent was obtained from the participant included in the study.

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Conflicts of interest

The authors declare that there are no conflicts of interests.

Data and materials availability

All data associated with this study are present in the paper.

REFERENCES AND NOTES

- Atalabi O, Ibinaiye O, Obajimi M, Ogunseyinde O, Akang E. Malignant haemangiopericytoma of the left thigh with metastases to the bones and lungs: A case report. *Nigerian J Surg Res* 2010; 8(3-4). doi:10.4314/njsr.v8i3-4.54899
- Benkirane S, BayBay H, Bennani M, Oulehri A, Mernissi FZ. Nodular Hidradenoma Case of Rare Localization. *Asp Biomed Clin Case Rep* 2020; 3(1):18-21 DOI: 10.36502/2020/ASJBCCR.6179
- Espat NJ, Lewis JJ, Leung D, Woodruff JM, Antonescu CR, Shia J. Conventional hemangiopericytoma: Modern analysis of outcome. *Cancer* 2002; 95(8):1746-51. doi: 10.1002/cncr.10867
- Fritchie K, Jensch K, Moskalev EA, Caron A, Jenkins S, Link M. The impact of histopathology and NAB2-STAT6 fusion subtype in classification and grading of meningeal solitary fibrous tumor/hemangiopericytoma. *Acta Neuropathol* 2019; 137(2):307-19. DOI: 10.1007/s00401-018-1952-6
- Grant EG, Grønvall S, Sarosi TE, Borts FT, Holm HH, Schellinger D. Sonographic findings in four cases of hemangiopericytoma. Correlation with computed tomographic, angiographic, and pathologic findings. *Radiol* 1982; 142(2):447-51. doi:10.1016/S0002-9394(14)70904-3
- Karcioglu ZA, Nasr AM, Haik BG. Orbital Hemangiopericytoma: Clinical and Morphologic Features. *Am J Ophthalmol* 1997; 124(5):661-72. doi:10.1016/S0002-9394(14)70904-3
- Mane K, Khan S, Singhania S, Singh PK, Gudhe M, Nikose S. Haemangiopericytoma of Left Thigh: A Rare Case Report. *J Orthopaed Rehabil* 2015; 1(2): 28-30
- Shruthi Madhavi G. Nodular Hidradenoma: A Forgotten Tumor of the Scalp. *J Dermatol Res Ther* 2021; 7(1). doi: 10.23937/2469-5750/1510095

9. Stout AP, Murray MR. Hemangiopericytoma a vascular tumor featuring zimmermann's pericytes: Ann Surg 1942; 116(1):26–33.
10. Tauro LF, George C, Kamath A, Sundaran S, Gatty R. Hemangiopericytoma of the Hand. Indian J Surg Oncol 2013; 4(1):76–9. doi: 10.1007/s13193-012-0194-7
11. Taylor KJ, Ramos I, Carter D, Morse SS, Snower D, Fortune K. Correlation of Doppler US tumor signals with neovascular morphologic features. Radiol 1988; 166(1):57–62. doi: 10.1148/radiology.166.1.2447604